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Slow-Growing Solitary Bulla of the Upper Arm

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Clinical Case

A 13 year-old Caucasian female presented with a slow-growing, solitary bulla on her left upper arm. The bulla first presented one year ago as a pin-sized tan papule. The papule enlarged slowly over one year to its current size of 1.5 cm x 1 cm and now a fluid-filled bulla with a firm nodular core. The bulla was not painful but mildly tender when manipulated. The patient was otherwise healthy and without any personal or family history of similar lesions.

On exam, there was a solitary, transluminating, pink, 1.5cm x 1cm fluid-filled bulla with a solid, nodular core on the left lateral upper arm. [Figure 1] There's a palpable dermal component of the lesion. The overlying skin was normal without telangiectasia, scale, induration, or inflammation. The surrounding skin was characterized by moderate keratosis pilaris. There was no known trauma to the site of lesion. An excisional biopsy of the lesion was performed and gross examination of the specimen revealed a flaccid bulla filled with serous fluid and a white, firm, calcified core. [Figure 2]

What is the diagnosis?

- A. Bullous morphea
- B. Secondary anetoderma
- C. Bullous pilomatrixoma
- D. Lymphangioma

Diagnosis

- C. Bullous pilomatrixoma

The lesion was excised with a narrow margin of 1mm given its benign clinical behavior: slow growth, no ulceration, and absence of associated systemic findings. Histopathology of the specimen revealed abundant clusters of eosinophilic anucleate “ghost cells” as well as large islands of basoloid cells [Figure 3 & 4] Interestingly, some of the basaloid islands demonstrated a high number (8-10) of mitotic figures per high power field, but given the overall well-circumscribed architecture and the lack of other concerning features, the final pathology was consistent with a bullous pilomatrixoma, a rare variant

of a common cutaneous nodule. The excisional surgery was uncomplicated and the patient healed well over the following weeks.

Discussion

While the other listed choices may present as a blister or bulla, the nodular core is a unique feature of bullous pilomatrixoma. Pilomatrixoma is a common type of benign adnexal tumor that arises from hair matrix cells and are associated with *CTNNB1* mutation, a gene that codes for β -catenin.¹⁻³ Bullous pilomatrixoma, also referred to as anetodermic pilomatrixoma or lymphangiectatic pilomatrixoma, is an uncommon variant of pilomatrixoma. In a recent study by Li, *et al*, bullous pilomatrixoma is found to share molecular features with classic pilomatrixoma as a result of the *CTNNB1* mutation.² Although no clinical syndromes have been linked to bullous pilomatrixoma, traditional pilomatrixoma has been associated with Rubinstein-Taybi syndrome, myotonic dystrophy, Gardner's syndrome, and Turner's syndrome.⁴⁻⁸ Given the clinical, histological, and genetic resemblance between bullous pilomatrixoma and traditional pilomatrixoma, it is reasonable to bear in mind these syndromic associations when evaluating patients with bullous pilomatrixoma.

Histologically, lymphatic congestion, dilation, and resultant lymphedema have been highlighted in the majority of reported cases. Other characteristic histology findings of bullous pilomatrixoma include well-differentiated eosinophilic anucleate shadow cells ("ghost cells") and clusters of basoloid cells, both of which were seen in our patient's specimen.² Among reported cases of bullous pilomatrixoma, the most common site of lesion is the upper arm.⁹⁻¹⁰ Based on a literature review by Chen, *et al*, bullous pilomatrixoma favors a female predominance and patients between ages of 10-20 years.¹⁰ The goal of this case report is to encourage providers to include bullous pilomatrixoma in the differential diagnosis for benign nodular lesions, and it's worth noting that our patient had a classic presentation for bullous pilomatrixoma based on current understanding of the condition.

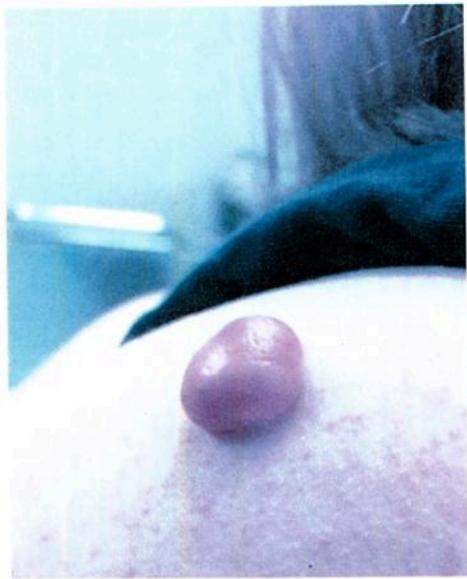


Fig. 1 Pink, 1.5cm x 1cm, fluid-filled bulla on the left lateral upper arm with background keratosis pilaris.

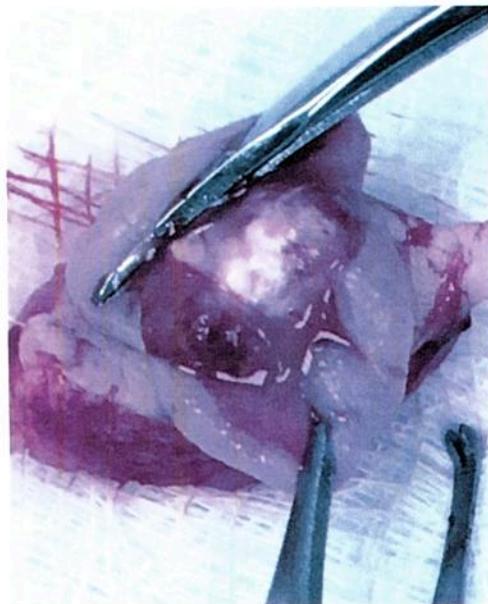


Fig. 2 Gross exam reveals a flaccid bulla filled with serous fluid and a white, firm, calcified core.

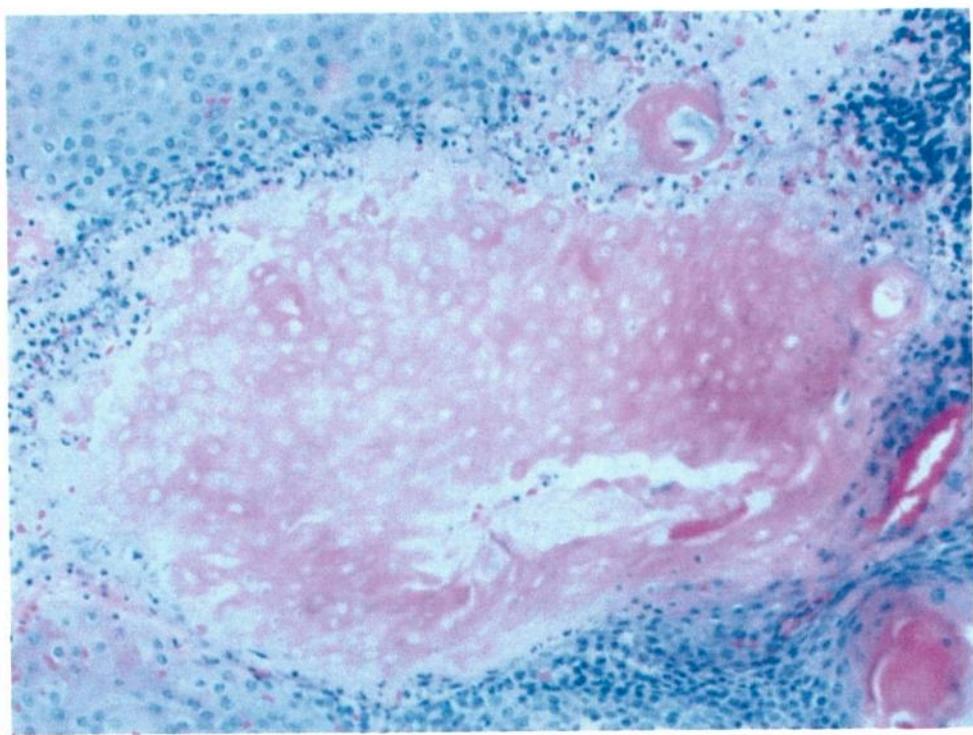


Fig. 3 H&E stain, 20x. Eosinophilic, anucleated shadow cells.

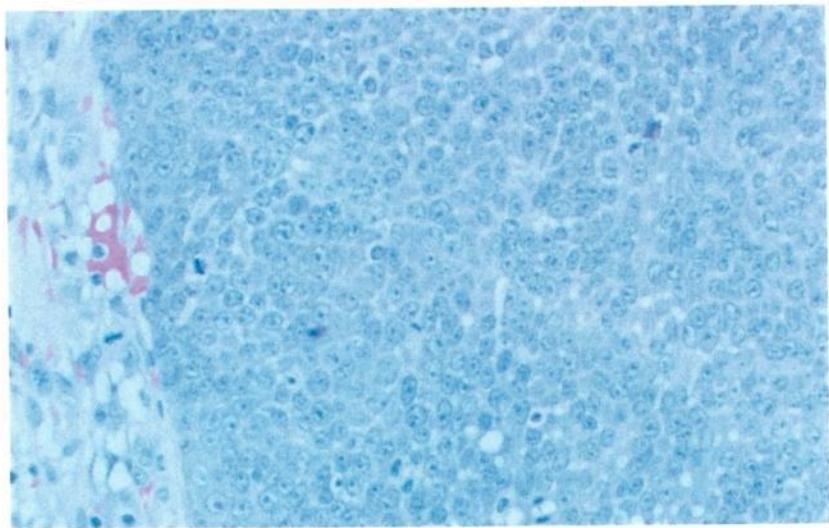


Fig. 4 H&E stain, 40x. Island of basaloid cells.

References

¹ Chmara M, Wernstedt A, Wasag B, et al. Multiple pilomatricomas with somatic CTNNB1 mutations in children with constitutive mismatch repair deficiency. *Genes, Chromosomes and Cancer* 2013;52:656–64.

² Li L, Zeng Y, Fang K, et al. Anetodermic pilomatrixoma: Molecular characteristics and trauma in the development of its bullous appearance. *Am J Dermatopathol* 2012;34:e41-5.

³ McCalmont TH. Adnexal neoplasms. In: Bolognia JL, Jorizzo JL, Rapini RP, editors. *Dermatology*. 2nd ed. Gurgaon: Mosby; 2009. pp. 1698–9.

⁴ Cambiaghi S, Ermacora E, Brusasco A, Canzi L, Caputo R. Multiple pilomatricomas in Rubinstein-Taybi syndrome: A case report. *Pediatr Dermatol* 1994;11:21–5.

⁵ Chiaramonti A, Gilgor RS. Pilomatrixomas associated with myotonic dystrophy. *Arch Dermatol* 1978;114:1363–5.

⁶ Cooper PH, Fechner RE. Pilomatrixoma-like changes in the epidermal cysts of Gardner's syndrome. *J Am Acad Dermatol* 1983;8:639–44.

⁷ Noguchi H, Kayashima K, Nishiyama S, Ono T. Two cases of pilomatrixoma in Turner's syndrome. *Dermatology* 1999;199:338–40.

⁸ Belliappa P, Umashankar N, Raveendra L. Bullous Pilomatrixoma: A Rare Variant Resembling Bouncy Ball. *International Journal of Trichology* 2013;5:32–4.

⁹ Erdogan HK, Kaya Z, Aytop CD, Acer E. A rare variant of pilomatrixoma: pseudobullous pilomatrixoma. *Act Dermatovenerol Alp Pannonica Adriat* 2015;24:59-60.

¹⁰ Chen SY, Wu F, Qian Y, Zhu L, Tu YT, Huang CZ. Pilomatrixoma with bullous appearance: a case report and review of literature. *Int J Dermatol* 2011;50:615-8. □